Discussion.– La maladie de Thévenard correspond au type 1 des neuropathies héréditaires sensitives et autonomes, de transmission autosomique dominante, qui sont beaucoup plus rares que les neuropathies de Charcot-Marie-Tooth. Elle évolue lentement à partir des deuxième et troisième décennies sous la forme de troubles de la sensibilité thermoalgésique, entraînant ainsi des ulcérations indolores au niveau des points de pression. Les ulcérations s'étendent et apparaissent alors des maux perforants plantaires, une dislocation du tarse avec un aspect de « pied cubique », des surinfections sous forme de panaris analgésiques, et enfin des altérations ostéoarticulaires du squelette du pied qui aboutissent à des mutilations des phalanges.
L'électroneuromyographique confirme la polyneuropathie essentiellement sensitive, la biopsie neuromusculaire permet d'écarter les autres polyneuropathies entraînant une altération de la sensibilité thermoalgique, telles les neuropathies diabétique, amyloïde, para-amyloïde et lépreuse. L'étude génétique confirme le diagnostic en mettant en évidence une mutation du gène *SPTLC1*.
Sur le plan thérapeutique, aucun traitement curatif n'est disponible. Toutefois, le traitement préventif des lésions cutanées est primordial.

http://dx.doi.org/10.1016/j.rehab.2012.07.528

P122-f

Estimation du score de handicap dans la sclérose en plaques (SEP) par l'analyse des données posturographiques

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Mots clés : Sclérose en plaques ; Posturographie ; Analyse quantifiée des récurrences
Introduction.– La SEP est une pathologie neurologique fréquente de l'adulte jeune et les troubles d'équilibre sont précoces. La posturographie est un moyen d'évaluation des troubles de l'équilibre en conditions orthostatiques. L'échelle d'évaluation du handicap utilisée habituellement dans la SEP est l'EDSS. Nous proposons une méthode d'estimation du score EDSS à partir des données posturographiques.
Matériel et méthode.– Cent dix-huit sujets sont inclus dans cette étude : 89 SEP définis selon MacDonald [1] : (âge = 46,1 ± 10,2, durée m = 13,4 ± 8,8 ans, EDSS = 2,74 ± 1) et 29 témoins (âge = 34,7 ± 12,3 ans). La posturographie est réalisée sur plate-forme Satel en conditions standardisées (t = 51,2 sec, Fq = 40 hz).
Nous avons effectué différentes mesures du centre de pression (CP) : longueur, surface, et analyse quantifiée des récurrences (RQA) [2], établies à partir des données posturographiques. Les coefficients de corrélation entre l'EDSS et chaque mesure RQA ont été mesurés. Une régression polynomiale de second ordre a été utilisée afin d'obtenir une estimation de l'EDSS. Enfin, nous avons calculé l'erreur moyenne entre score estimé et score observé.
Résultats.– Les meilleures corrélations au score EDSS sont observées avec l'entropie (R = 0,8448, $p < 0,05$). Les meilleurs résultats entre EDSS observé et EDSS calculé à partir des données posturographiques sont obtenus grâce au pourcentage de récurrence (70,49 %, erreur moyenne EDSS = 0,63).
Discussion/conclusion.– Nous avons étudié plusieurs méthodes de calcul du score EDSS à partir des données posturographiques. Les accélérations et vitesses semblent moins discriminantes que les autres valeurs RQA. La vitesse et l'accélération croissent proportionnellement au score EDSS (Anova, $p < 0,05$), sauf pour EDSS = 4 (erreur moyenne plus importante). Dans cette étude nous montrons qu'il est possible d'estimer le score EDSS à partir des données posturographiques. Il serait intéressant de combiner cette méthode à d'autres mesures afin d'établir une modélisation de la régulation de l'équilibre dans la SEP.

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http://dx.doi.org/10.1016/j.rehab.2012.07.529

English version

P118-e

Progression of scoliosis after intrathecal baclofen in an adult patient with multiple sclerosis

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Keywords: Intrathecal baclofen; Scoliosis; Multiple sclerosis
Introduction.– Intrathecal baclofen (ITB) induces modification of axial tonus. The role of ITB in the evolution of scoliosis has been studied mainly in cerebral palsy. We present the case report of an adult female multiple sclerosis patient who developed a major scoliosis after implantation of a baclofen pump.
Case report.– A woman, 45 years old, with spastic quadriplegia secondary to multiple sclerosis evolving since 1984 presents thoracolumbar pain 30 months after intrathecal baclofen pump insertion in 2006 at the age of 40. Plain X-rays show a rapidly progressing right thoracolumbar scoliotic curve (Cobb angle 15/10/2008: 7° versus 54° on the 12/01/12) requiring a T3-sacrum posterior fusion (01/02/2012). This patient has no history of idiopathic scoliosis and had not reported backpain prior to implantation.
Discussion.– Several studies report the evolution of scoliosis after BIT in cerebral palsy and discuss the role of the treatment versus the evolution due to skeletal maturation [1,2]. Our case report highlights that major aggravation of scoliosis may occur after ITB, in adults outside the risk periods of adolescence and menopause. This draws our attention to the necessity of regular follow up of the spine after ITB.

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http://dx.doi.org/10.1016/j.rehab.2012.07.530

P119-e

Neuro-vitamin B12 deficiency: 25 cases and experience of the Ben Aknoun rehabilitation unit, Algeria

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Introduction.– Neurological disorders may develop in patients with vitamin B12 deficiency. The predominant sign is combined spinal cord sclerosis which may be invalidating, even after vitamin replacement therapy.
Material and method.– We report a retrospective study of 25 patients treated in the PRM unit of the Ben Aknoun Hospital in Algiers during period 2000 to 2010.
Results.– The series included 25 patients: 14 women and 11 men, mean age 43.96 years. Patients were were mainly referred from neurology ($n = 20$ patients). Biermer's anaemia predominated. The inaugural signs of the disease

were essentially: gait disorders (16 patients) and paresthesia (four other patients). The diagnosis of vitamin deficiency B12 and the implementation of the treatment were relatively late, on average 14 months after symptom onset. The physical examination found a posterior cord syndrome associated with a pyramidal syndrome in all the patients. It is noteworthy that a cerebellar syndrome was found in three patients and a vestibulary syndrome in two.
The functional assessment found:
– eight wheelchair patients;
– 11 patients used technical aids for walking;
– six patients walked without help.
Care included rehabilitation sessions for all patients; eight patients required or anti-spasticity treatment. The outcome was marked by a relative functional improvement in 22 patients; three patients still had to use the wheelchair.
Conclusion.– The functional recovery of patients presenting combined spinal cord sclerosis depends on the gravity of the clinical deficiencies, on the precocity of the diagnosis and early treatment with vitamin replacement therapy.

http://dx.doi.org/10.1016/j.rehab.2012.07.531

P120-e

Tiredness and sequelae poliomyelitis

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Keywords: Tire; Syndrome post-polio; Multidisciplinary treatment
Objectives.– Tiredness is frequently met among survivors of poliomyelitis. The objectives of our study is to find the incidence of tiredness among our surviving Algerian patients of poliomyelitis, and to seek the correlation between the degree of tiredness and the various biometric and sociodemographic parameters.
Materials and methods.– A descriptive and prospective clinical study of 74 patients surviving of poliomyelitis, seen in consultation between years 2009 and 2012, by using a drawn up card taking in count the variables of balance of the various parameters: tiredness, biometric data, socio professional data and the various clinical signs of the syndrome post-poliomyelitis. SPPS 14.0 software used for the epidemiologic study.
Results.– The incidence of tiredness among survivors of poliomyelitis is considerably present at 80%, the peak of age between 40 to 50 years, the mailmen biometric (age, weight and IMC) and socioprofessional does not seem to have a significant influence on tiredness. A percentage of 70.3 of the survivors of polio present a syndrome post-poliomyelitis, and only the amyotrophie is found in significant report/ratio ($P < 0.03$).
Conclusions.– The assumption of responsibility must be based on the origin and the diagnosis of tiredness.
The treatment of tiredness must consist of an education of the patient on the syndrome post-polio, the effort economy, the management of the rest, and the recourse to technical assistances. Rehabilitation to this end should act within a multidisciplinary framework.

http://dx.doi.org/10.1016/j.rehab.2012.07.532

P121-e

Thevenard's disease or ulcero-mutilating acropathology syndrom: Case report and literature review

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Keywords: Thevenard's disease; Ulcero-mutilating acropathology
Introduction.– The Thevenard disease is a rare familial ulcero-mutilating acropathology, responsible for sensory peripheral neuropathy associated with dysautonomic syndrome. In clinical practice, diagnosis relies on the clinical data, electrophysiological and family. It is confirmed by molecular biology, neuromuscular biopsy an interest in differential diagnosis. The objective of this work and to recall this rare disease, often a source of handicap and difficulties in diagnosing and management.
Observation.– Mr. BM, 77 years old, who presented a peripheral neuropathy hereditary sensory and dysautonomic evolving since the age of 15 years and complicated plantar ulcers in both feet and repetitions of a chronic osteomyelitis of the metatarsophalangeal joints left. The electroneuromyography (EMG) showed a peripheral sensory polyneuropathy. Neuromuscular biopsy lead to Wallerian degeneration associated with hypomyelination. Radiographs of the left foot showed lysis of tarsus and metatarsus bones, leading to transmetatarsal amputation. A directed healing and an apparatus using initially a transitional aid to healing of the foot and orthopedic shoes are allowed for the recovery of autonomy in walking.
Discussion.– Thevenard's disease refers to the type 1 hereditary neuropathies, sensory and autonomous autosomal dominant, who are much rarer than the Charcot-Marie-Tooth neuropathy. It evolves slowly from the second and third decades in the form of sensory disturbances thermoalgesic, causing painless ulcers at pressure points. The ulcerations extend and appear then the plantar ulcers, a dislocation of the tarsus with an aspect of ''cubic foot'', secondary infections in the form of analgesic panaris, and finally osteoarticular alterations of the skeleton of the foot that result in mutilation of the phalanges.The electroneuromyography (EMG) confirms the predominantly sensory polyneuropathy, neuromuscular biopsy can rule out other polyneuropathies resulting in altered sensitivity thermoalgesic, such as diabetic neuropathy, amyloid and paraamyloid neuropathy and leprous neuropathy. Genetic study confirms the diagnosis by identifying a mutation in *SPTLC1*.On the therapeutic level, no cure is available. However, preventive treatment of skin lesions is promordial.

http://dx.doi.org/10.1016/j.rehab.2012.07.533

P122-e

Estimated score of disability in multiple sclerosis (MS) through posturographic data analysis

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Keywords: Multiple sclerosis; Posturography; Recurrence quantified analysis
Introduction.– MS is a common neurological disease in young adults and balance disorders appear early. Posturography is an evaluation of balance disorders in orthostatic conditions. The scale used for assessing disability in MS is usually the EDSS. We propose a method for estimating the EDSS from posturographic data.
Material and method.– One hundred and eighteen subjects were included in this study: 89 MS defined by a MacDonald [1] (age = 46.1 ± 10.2 years, dd = 13.4 ± 8.8 years, EDSS = 2.74 ± 1) and 29 controls (age = 34.7 ± 12.3 years). Posturography was performed on Satel platform in standardized conditions (t = 51.2 sec, Fq = 40 Hz). We carried out various measures of center of pressure (CP): length, area, and recurrence quantified analysis (RQA) [2], established from posturographic data. Correlation coefficients between the EDSS and each RQA measurement were performed. A second order polynomial regression was used to obtain an estimation of the EDSS. Finally we calculated the average error between observed and estimated score.
Results.– The best correlations with EDSS were observed with the entropy ($R = 0.8448$, $P < 0.05$). The best results between observed and calculated EDSS from posturographic data were obtained through the recurrence percentage (70.49%, average error EDSS = 0.63).